

Case report of a rare cause of intestinal obstruction: Abdominal cocoon syndrome

It is rare but all surgeons should know

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Abstract

Abdominal cocoon is a rare cause of ileus characterized by a pouch of a cocoon-like thick fibrous membrane partially or completely covering the small intestine and presents with acute and subacute intestinal obstruction findings. In this paper, we aimed to present a case operated for intestinal obstruction due to abdominal cocoon syndrome in light of the literature.

Keywords

Abdominal Cocoon; Intestinal obstruction; Peritoneal fibrosis; Ileus

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Introduction

Abdominal cocoon syndrome (idiopathic sclerosing capsule peritonitis) is a rare cause of intestinal obstruction [1]. Its most common presentation is acute or chronic small intestinal obstruction [2]. Although the etiology of the disease is not clearly known, some etiological factors include congenital/idiopathic or secondary sarcoidosis, tuberculosis, peritonitis, beta-blocker use, post-chemotherapy, peritoneal dialysis, post-transplantation, post-parasitic infections, post-trauma and previous abdominal surgery [3, 4]. Frequent radiological imaging is normal and intestinal segments covered in a membrane can be seen as a helix mark on computed tomography (CT) [5]. Definitive diagnosis is made in the light of intraoperative and histopathological findings.

Case Report

A 42-year-old male patient was admitted to the emergency department with complaints of loss of appetite, nausea, abdominal pain, inability to pas gas-stool and bloating in the abdomen and was admitted to the general surgery clinic with the preliminary diagnosis of intestinal obstruction. The patient had complaints of increasing bloating after meals for about 3 years.

The patient's abdominal examination revealed marked distention, sensitivity in all quadrants of the abdomen, defense, and rebound.

Laboratory Examination

Routine blood test results were as follows: Glucose: 203 mg/dL, LDH: 356, WBC: 15950/mm3 , no biochemical abnormal value except CRP: 20.4 mg/L.

Imaging

In the abdominal CT imaging of the patient, a cluster formation in the small intestine loops starting from the epigastric region to the paraumbilical region, thickening in the cocoon-like peritoneum surrounding it and an appearance of a fibrosis tissue are observed on the left quadrant of the abdomen. The appearance conforms with abdominal cocoon syndrome. Localized fluid accumulation is observed in the left subdiaphragmatic distance. The fluid accumulation conforms with the anterior superior of the defined abdominal cocoon contour. Its size was 85x35mm.

The patient's oral intake was discontinued; nasogastric decompression and IV fluid support were given. After a 24-hour follow-up, there was no regression in the patient's abdominal pain and physical examination findings, and the patient was operated under emergency conditions with a diagnosis of acute abdomen. It was difficult to enter the abdomen during the operation. It was observed that the peritoneum widely thickened. When the abdomen was entered, exploration revealed all small intestines to be wrapped by a gato-shaped lobule fibrous membrane and obstructed within the capsule towards the left upper quadrant and lower quadrant (Figure 1-2). Encapsulated and thickened fibrous bands on all small intestines were excised to maintain the integrity of the small intestine wall (Figure 3). The patient had oral and gas-stool discharge during the postoperative follow-ups and was discharged without any complications.

Histopathological examination

Pathology specimen consisted of macroscopically gray-white fibrotic membranous structures and yellow-colored fibroadipose tissue fragments with a limited amount of bleeding at the edge. Microscopically, fibrotic membranous structure, fibroblast and collagen fiber proliferation, hemorrhage, edema, fibrin exudation, chronic inflammatory cells, and fibrocollogenous tissue fragments were observed (Figure 4-5-6).



Figure 1. Abdominal CT: A cluster formation in the small intestine loops starting from the epigastric region to the paraumbilical region, thickening in the cocoon-like peritoneum surrounding it and an appearance of a fibrosis tissue are observed on the left quadrant of the abdomen. The appearance conforms with abdominal cocoon syndrome

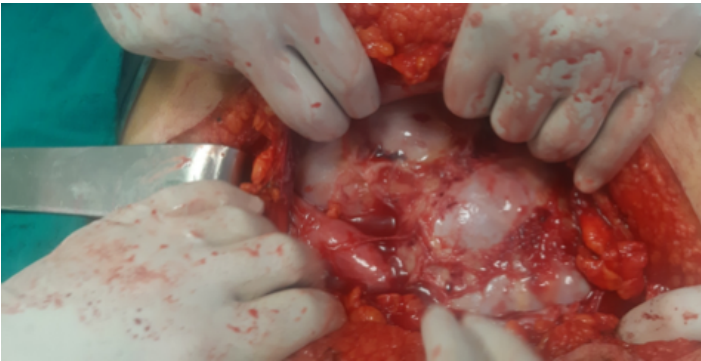


Figure 2. All small intestines to be wrapped by a gato-shaped lobule fibrous membrane and obstructed within the capsule towards the left upper quadrant and lower quadrant.



Figure 3. Encapsulated and thickened fibrous bands on all small intestines were excised to maintain the integrity of the small intestine wall.

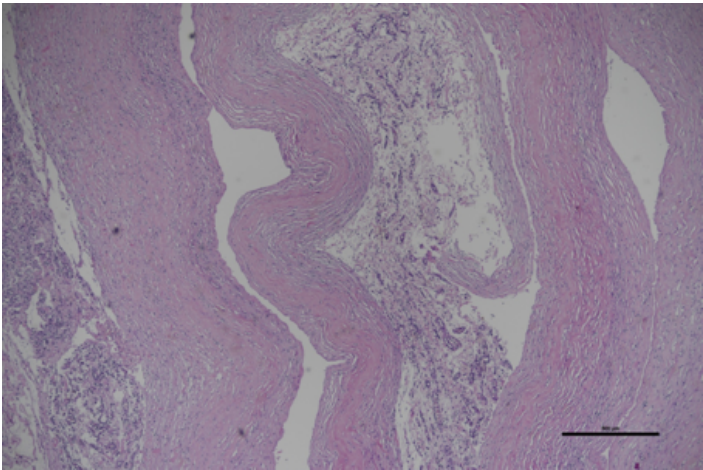


Figure 4. Fibrotic membranous structure, fibrocollagenous tissue fragments, chronic inflammatory cells, ectatic vascular structures (Hematoxylin Eosin, X40) (From Dr. Havva Hande KESER ŞAHİN's archive)

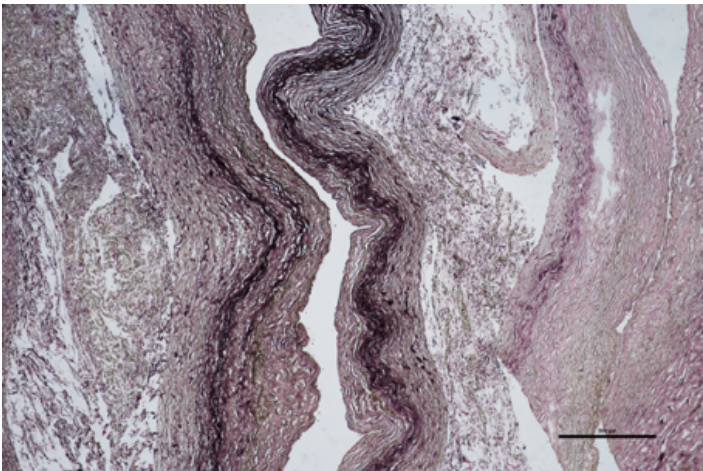


Figure 5. Proliferation in collagen fibers (Elastic Van Gieson, X40) (From Dr. Havva Hande KESER ŞAHİN's archive)

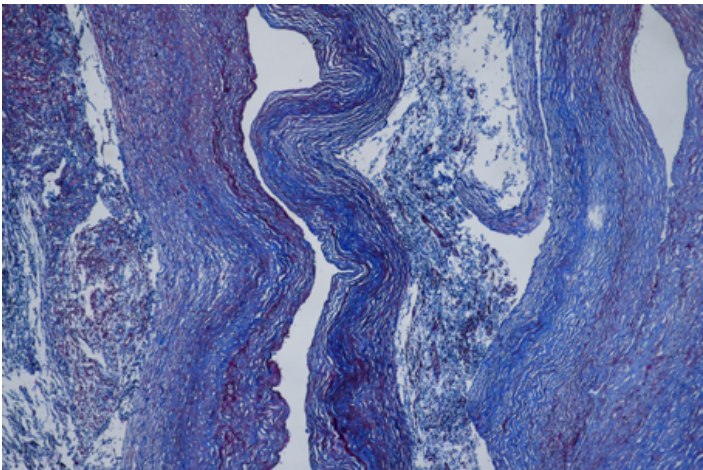


Figure 6. Proliferation in collagen fibers (Masson Trichrome, X40) (From Dr. Havva Hande KESER ŞAHİN's archive)

Discussion

Abdominal cocoon syndrome (ACS), also known as idiopathic sclerosing capsule peritonitis, is a rare cause of ileus. When PubMed is researched, it is seen that fewer than 150 cases have been reported to date [6]. It was first reported by Foo et al. [2]. ACS is a rare peritoneal disease with unclear pathogenesis. ACS can be divided into two, as idiopathic and secondary [7]. It has been reported that the idiopathic form may be associated with congenital dysplasia [8]. Many causes that may lead to peritonitis as a secondary etiology have been mentioned before. Our case was evaluated as idiopathic etiology considering that it could be a congenital condition due to the absence of a previous abdominal surgery and the complaints of abdominal pain after meals and no secondary pathology. The clinical appearance of ACS often appears as acute abdominal pain. In our case, it was determined that there was chronic pain in addition to acute abdominal pain. In addition to abdominal pain, symptoms such as nausea, vomiting, inability to pass gas-stool, as well as increased abdominal pain and a mass found by hand in physical examination, which may be signs of peritonitis, may be encountered [9]. Clinical signs and symptoms may vary depending on the severity and duration of the disease, the underlying causes and the patient's immunological status. ACS may be asymptomatic for years and suddenly manifest itself with abdominal pain and ileus findings, which has been associated with a change in lifestyle [9]. This has been explained as follows: when fatty and heavy dishes are consumed, the small intestine peristalsis increases significantly, and as a result, since the fibrous membrane covering the small intestine in ACS cannot be dilated, the small intestines get obstructed, and symptoms such as abdominal pain occur [9]. In our case, 3 days prior to admission to the hospital, the patient had a fatty meal more than a normal amount. Since there is no characteristic clinical finding, it is often difficult to diagnose ACS in the preoperative period. Barium contrast, X-ray, and high-resolution magnetic resonance imaging (MRI) or computed tomography (CT) can be helpful in the preoperative diagnosis of ACS [10]. Abdominal X-ray findings are not specific and multiple levels of air fluid can be seen, and characteristics of intestinal obstruction such as dilated loops of the small intestine and sometimes intestinal wall and peritoneal calcification can be detected [3, 11, 12]. Contrast-enhanced CT is the most reliable test to diagnose ACS. One CT finding is a cocoon-like surrounding of the small intestines by a dense membrane with non-contrasted boundaries. According to the study by Candido et al., it was found that peritoneal thickening is 100% detectable, the local collection is 90% detectable, calcification is 70% detectable, small intestinal loops in the middle of the abdomen are 60% detectable and peritoneal enlargement is 50% detectable with CT in ACS [12]. In our case, the preoperative pre-diagnosis was made with the help of an abdominal X-ray and especially CT. Surgery is considered the gold standard treatment for ACS [13]. Surgery is often long and difficult, and recurrence of intestinal obstruction due to intestinal adhesion may occur after surgery. In asymptomatic or minimally symptomatic cases, if the diagnosis can be made with CT, it can be managed with conservative treatment [9]. In our case, the patient was first

given conservative treatment. However, since there was no decrease in the laboratory and physical examination findings of the patient, a surgical decision was made.

Pathological examination of the surgically excised peritoneal membrane will show interstitial thickening consisting of fibroblasts and collagen accumulation in the peritoneal membrane but a complete loss in the mesothelium [14]. Inflammatory cells are always present, but leukocyte infiltration is not a must for diagnosis [14].

In the pathogenesis of cocoon formation, It is thought that fibrogenic cytokines are released from fibroblasts trigger neoangiogenesis and peritoneal fibrosis [15, 16].

In conclusion, ACS is a rare cause of acute abdomen that is difficult to diagnose preoperatively. It is a clinical entity that we may have to consider in a patient with signs of intestinal obstruction even when there is no unexplained abdominal pain, abdominal mass or previous abdominal surgery. Its etiology and pathogenesis remain uncertain, and its clinical presentation is not specific. Clinical signs of obstruction may be observed as aggressive or limited. Thus, we suggest making the decision for surgery based on the patient. Ultimately, the purpose of surgery is to eliminate the obstruction, but it may not be possible to excise the whole membrane, which should not be the aim. In asymptomatic patients, a healthy diet and lifestyle training should be given and applied to avoid the onset of acute ACS.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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